Session 4: Neurodegenerative Neuropathology

Abstract 9

Chronic traumatic encephalopathy (CTE) is absent from a European community-based aging cohort while cortical aging-related tau astrogliopathy (ARTAG) is highly prevalent

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Chronic traumatic encephalopathy (CTE) and aging-related tau astrogliopathy (ARTAG) are characterised by tau-immunopositive neuronal and/or astrocytic inclusions, with overlapping cortical involvement and astrocytic inclusion morphology. This study determined the prevalence of CTE and cortical ARTAG in a European community-based population (n=310) and explored overlap of both pathological entities. Frontal, parietal and temporal cortices were assessed. No case fulfilling CTE criteria was found. However, isolated astroglial or neuronal tau pathologies were recognized in sulcal depths (<2%). One case without history of traumatic brain injury showed combined tau-immunoreactive features confined to frontal sulci without perivascular accumulation. Another 24 cases had single tau pathologies in cortical sulci. ARTAG was identified in 117 cases (38%), with a similar regional prevalence. Grey matter ARTAG was the most common followed by subpial, white matter and perivascular. The presence of any type of ARTAG was associated with having another type of ARTAG in the same region ($P<0.05$). In summary, cortical ARTAG in this population is common and contrasts the high prevalence of CTE in individuals with repeated mild traumatic brain injury.

LEARNING OBJECTIVES

This presentation will enable the learner to:
Classify tau-immunopositive astrocytic inclusions characteristic of ARTAG

1. Describe neuropathological components of CTE
2. Identify CTE and cortical ARTAG in a case series

Abstract 10

Nodding syndrome, an epidemic young-onset epilepsy-dementia complex in Uganda

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Nodding syndrome (NS) is an enigmatic recurrent epidemic neurological disease that affects children in East Africa. The illness begins with nodding of the head and grand mal seizures that may lead to death after several years. The most recent outbreaks of NS occurred in northern Uganda and South Sudan. We describe the clinicopathologic spectrum of NS in Uganda. Ten children or young adults with NS were studied at autopsy and the neuropathological findings correlated with the onset, duration and progression of their neurological illness. All cases had epilepsy with grand mal seizures. Three cases had a clinical course that was predominantly characterized by epilepsy. Seven patients had progressive frontotemporal dementia. Two of the patients with dementia also had progressive quadriparesis. In all cases, the brain revealed tau pathology. In cases with an epilepsy-predominate course, the tau pathology was largely limited to the anterior frontal lobes but cases with dementia had more widespread cortical and subcortical tau pathology. In some cases, the histologic pattern was reminiscent of progressive supranuclear palsy. There are some interesting parallels between NS and the amyotrophic lateral sclerosis/Parkinson-dementia complex (ALS/PDC). The similarities are the presence of geographical isolates of disease manifesting in indigenous populations with familial clusters but no clear heritability. Both disorders appear to be related to an unknown environmental factor and both diseases appear to be fading over time in the respective geographical locations. One of the major open questions is whether ALS occurs in NS. This question will be addressed in future clinical studies and postmortem examination of the spinal cord. We propose that NS is a unique epilepsy-dementia complex in East Africa.

LEARNING OBJECTIVES

This presentation will enable the learner to:

1. Describe the clinicopathologic features of a nodding syndrome.
2. Compare the pathology of NS to ALS/PDC and related disease

Abstract 12

The Amygdala in Neurodegeneration

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The amygdala is a key anatomic structure that has multiple different nuclei and is involved in several critical aspects of cognition and systemic functions. Several different neurodegenerative diseases have major pathological effects on distinct amygdala nuclei. This presentation will describe the classic and characteristic anatomic distributions in the amygdala of "pure" Alzheimer disease and "pure" Lewy body disease, as well as "normal aging". In addition, data will be presented on how these classic distributions are altered in either "mixed dementias" or in some atypical forms of neurodegeneration. Amygdala pathology will also be illustrated in several other neurodegenerative diseases. The implications of the differing anatomic distributions in different neurodegenerative diseases will be discussed.